DOI: 10.7860/JCDR/2025/78831.21537

Case Report

Pathology Section

A Rare Paediatric Case of Double Appendix Anomaly with Appendiceal Neuroendocrine Neoplasm

KOVURI UMADEVI[1], BHAVANI SHANKER NITYANANDA[2], ZUAFSHAN SULTANA[3], MAHENDRA J PARAGE[4]

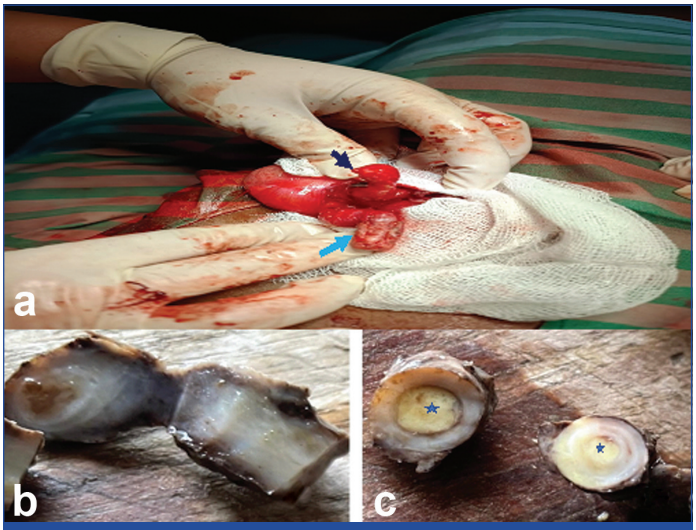
ABSTRACT

This case report presents a rare congenital anomaly: the presence of a double appendix, a condition observed in approximately 1 in 25,000 appendectomy cases. The discovery of a neuroendocrine tumour in one of the appendices further underscores the exceptional nature of this case. Present case is of an eight-year-old boy who was admitted with acute abdominal pain, during which emergency surgery revealed a double appendix. While one appendix appeared normal, the other exhibited a yellow nodular lesion suggestive of malignancy. Histopathological evaluation confirmed a Grade I well-differentiated neuroendocrine tumour measuring 1.2 cm. The tumour had invaded the serosa and the proximal surgical margin but showed no lymphovascular invasion. Immunohistochemical analysis demonstrated strong positivity for chromogranin and negativity for synaptophysin, with a low Ki-67 index of 3%, corroborating the diagnosis. Following National Comprehensive Cancer Network (NCCN) guidelines, the patient underwent a right hemicolectomy to ensure complete tumour removal and minimise the risk of recurrence. Subsequent histological examination showed no residual tumour or lymph node metastases. The patient recovered well, with no recurrence noted during follow-up. This case highlights the need for careful intraoperative exploration and thorough pathological assessment, particularly when rare anatomical anomalies are encountered. Documenting such unique findings enriches medical knowledge, raises clinical awareness and underscores the value of evidence-based surgical management in achieving optimal outcomes.

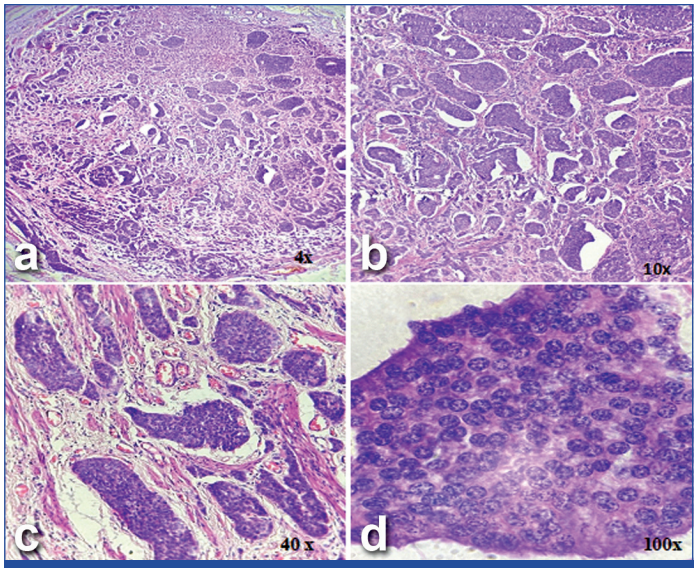
Keywords: Carcinoid tumour, Congenital abnormalities, Guideline adherence, Neuroendocrine tumours, Operative, Rare diseases, Right hemicolectomy, Surgical procedures, Treatment outcome

CASE REPORT

An eight-year-old male presented to Niloufer Hospital in Hyderabad with acute abdominal pain that was clinically suggestive of acute appendicitis. Emergency surgery was performed and intraoperatively, surgeons discovered a rare congenital anomaly—two distinct appendices originating from the caecum. One appendix appeared grossly normal, while the other showed a yellow nodular lesion, raising suspicion for neoplasia. Gross pathological examination confirmed the presence of a tumour in the second appendix [Table/Fig-1]. Histopathological examination revealed a Grade I well-differentiated neuroendocrine tumour (carcinoid tumour) measuring 1.2 cm in maximum diameter. The tumour displayed a well-circumscribed nodular growth pattern with lobulated margins and fibrous septation, surrounded by reactive desmoplastic stroma [Table/Fig-2].



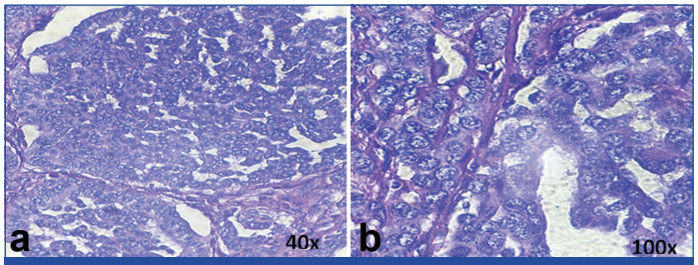
[Table/Fig-1]: a) Intraoperative findings revealed a rare duplex appendix anomaly, with two appendices arising from the caecum (blue arrows); b) One appendix appeared normal, with a patent lumen and no gross abnormalities; c) Second appendix contained a distinct yellowish lesion tumour-like lesion, well-demarcated and occupying the luminal space, denoted by a star.



[Table/Fig-2]: a) Well-circumscribed, lobulated tumour separated by fibrous septa with surrounding desmoplastic stroma (H&E, 4x); b) Tumour cells arranged in nests and trabeculae within reactive stroma (H&E, 10x); c) Uniform round nuclei with salt-and-pepper chromatin and moderate eosinophilic cytoplasm; no mitoses or atypia, tumour infiltration into the serosa is evident (H&E, 40x); d) Prominent nuclear detail with salt and pepper chromatin (H&E, 100x).

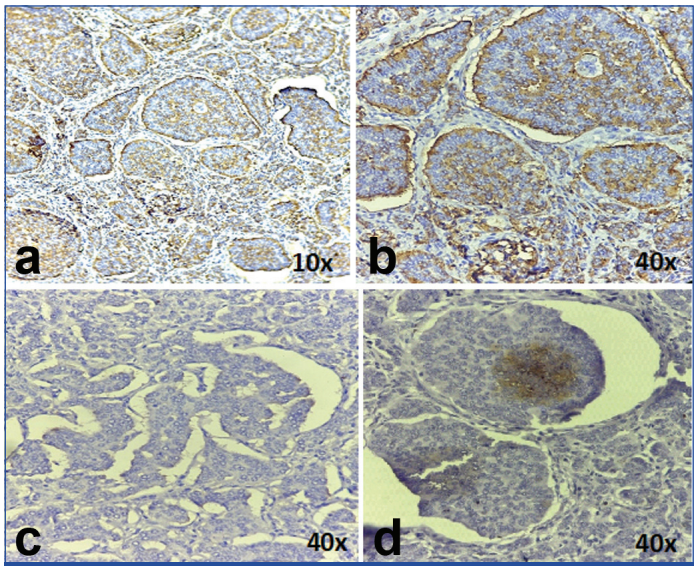
At 10x magnification, tumour cells were organised in nests and trabeculae with intervening thin fibrous septa. At 40x, the tumour cells showed round nuclei with finely stippled “salt-and-pepper” chromatin and moderate eosinophilic cytoplasm, with no significant mitotic activity or nuclear atypia. Oil immersion (100x) highlighted nuclear detail and amphophilic cytoplasm, confirming neuroendocrine morphology. Tumour invasion into the serosa

was evident. Periodic Acid-Schiff (PAS) staining provided valuable insights into the structural and cytoplasmic characteristics of the tumour. At low-power magnification (4x and 10x), the stain clearly delineated the tumour's lobulated architecture and well-demarcated basement membranes, helping to highlight the organised nature of the lesion. At higher magnifications (40x and 100x), PAS positivity was observed within the cytoplasm, reflecting the presence of glycogen—features that support neuroendocrine differentiation [Table/Fig-3].



[Table/Fig-3]: a) Tumour cells with stippled chromatin and cytoplasmic PAS positivity, indicating glycogen (PAS, 40x); b) Enhanced nuclear detail and glycogen-rich cytoplasm, supporting neuroendocrine differentiation and stromal invasion (PAS, 100x).

Immunohistochemistry confirmed neuroendocrine differentiation. Chromogranin A staining showed granular cytoplasmic positivity at both low and high magnifications [Table/Fig-4a,b], while synaptophysin exhibited diffuse cytoplasmic positivity [Table/Fig-4c]. Ki-67 immunostaining, evaluated at 40x magnification, demonstrated focal nuclear positivity with a labeling index of approximately 3%, classifying the tumour as a Grade 2 (intermediate-grade) neuroendocrine tumour according to the World Health Organisation (WHO) 2019 classification [Table/Fig-4d] [1]. Additional histopathological features included serosal invasion, proximal surgical margin involvement and perineural invasion, although no lymphovascular invasion was identified. Given the margin positivity and invasive features, the patient underwent a completion right hemicolectomy, in accordance with NCCN guidelines for the management of appendiceal neuroendocrine tumours with adverse prognostic indicators. Histopathological evaluation of the resected hemicolectomy specimen revealed no residual tumour and no evidence of lymph node metastasis. The patient's postoperative recovery was uneventful and he continues to remain recurrence-free on follow-up.



[Table/Fig-4]: a) Immunohistochemical analysis demonstrates strong granular cytoplasmic positivity for Chromogranin A at low magnification (10x), highlighting neuroendocrine differentiation; b) At higher magnification (40x), Chromogranin A staining reveals intense cytoplasmic positivity with well-preserved cell morphology; c). Synaptophysin negative; d) Ki-67 staining at 40x demonstrates focal nuclear positivity with an index of 3%, consistent with a low to intermediate-grade neuroendocrine tumour (WHO Grade 2).

DISCUSSION

A double appendix is an exceedingly rare congenital anomaly, with an incidence estimated at approximately 1 in 25,000 appendectomies [1]. This condition was first described by Picoli in 1892. It is generally classified into Type A (both appendices sharing a common base) and Type B (each appendix arising independently from the caecum) [2]. Most cases are detected incidentally during surgeries performed for unrelated abdominal conditions or at autopsy [3].

Appendiceal carcinoid tumours are rare neoplasms, accounting for less than 1% of all gastrointestinal neuroendocrine tumours [4]. They arise from neuroendocrine (Kultschitzky) cells and demonstrate both endocrine and neural differentiation [5,6]. These tumours are more commonly encountered in adults and paediatric presentations are rare [7]. The occurrence of a neuroendocrine tumour in one appendix of a double appendix configuration is exceedingly unusual, with very few reported cases worldwide [8].

This case is unique due to the co-existence of two rare pathologies: a double appendix and an incidental well-differentiated neuroendocrine tumour in a paediatric patient. The tumour size (1.2 cm), presence of subserosal invasion and proximal surgical margin involvement were critical findings guiding further management. According to established NCCN guidelines, tumours under 2 cm without high-risk features may be managed with appendectomy alone. However, the presence of adverse features, such as positive margins, perineural invasion, or mesoappendiceal extension, mandates more extensive resection [5,9].

In the present case, the revision right hemicolectomy was warranted due to the involvement of the proximal surgical margin. Histopathology confirmed complete resection with no residual tumour or lymph node involvement, indicating a favourable outcome. The patient's recovery was smooth and he remains under disease-free surveillance [10].

The embryological mechanism behind double appendix formation is not fully understood but is thought to involve errors during midgut rotation or duplication of caecal diverticula during the 5th to 8th weeks of foetal development [11]. In terms of the pathogenesis of neuroendocrine tumours, genetic and molecular factors such as MEN1 mutations and chromosomal losses have been postulated in adults, though these mechanisms remain speculative in paediatric populations [12].

The prognosis for well-differentiated appendiceal carcinoid tumours is generally excellent when diagnosed early and treated appropriately. Localised tumours have a five-year survival rate exceeding 90%, which decreases in the presence of nodal or distant metastases. For tumours with lymph node involvement, survival drops to about 84.6%, and in cases with distant metastasis, survival declines to 33.7% [13]. There are limited case reports describing appendiceal carcinoid tumours in the setting of a double appendix. Shibata J et al., reported a 69-year-old patient with a 1.5 cm carcinoid tumour successfully treated with appendectomy [11]. Another case reported by Coşkun H et al., described a 2 cm tumour in a 29-year-old, also managed with conservative surgery and yielding a favourable outcome [14].

The current case stands out due to the patient's young age and the need for a hemicolectomy, highlighting the importance of margin status and tumour behaviour in determining surgical strategy. The significance of recognising anatomical variants, such as a double appendix, lies in their diagnostic, therapeutic and medicolegal implications. Failure to identify a second appendix intraoperatively could lead to missed diagnoses, especially if the second appendix harbours pathology. Thus, comprehensive intraoperative assessment and meticulous pathological evaluation are essential.

CONCLUSION(S)

This case highlights the clinical importance of recognising and managing rare anatomical anomalies, such as a double appendix, especially when accompanied by an incidental neuroendocrine tumour. The favourable outcome in this paediatric patient was made possible through prompt surgical intervention, detailed histopathological evaluation and the application of standardised oncological guidelines. Documenting such rare associations contributes to the broader understanding of congenital variants and their potential clinical implications. It also emphasises the role of surgical awareness and multidisciplinary collaboration in ensuring accurate diagnosis, timely management and improved long-term outcomes in paediatric oncology.

REFERENCES

[1] Wang HY, Li ZW, Sun W, Yang X, Zhou LX, Huang XZ, et al. Automated quantification of Ki-67 index associates with pathologic grade of pulmonary neuroendocrine tumours. Chin Med J. 2019;132(05):551-61.

[2] Singh S, Chalotra V, Jain A, Pal Singh T. Double appendix: A rare case entity. Asian J Case Rep Surg. 2022;16(1):9-12. Available from: http://research.manuscritpub.com/id/eprint/863.

[3] Foorthuis R. On the nature and types of anomalies: A review of deviations in data. International journal of data science and analytics. Int J Data Sci Anal. 2021;12(4):297-331. Doi: 10.1007/s41060-021-00265-1. PMID: 34368422; PMCID: PMC8331998.

[4] Gu Y, Wang N, Xu H. Carcinoid tumour of the appendix: A case report. Oncol Lett. 2015;9(5):2401-03. Available from: https://doi.org/10.3892/ol.2015.2997.

[5] Nageswaran H, Khan U, Hill F, Maw A. Appendiceal duplication: A comprehensive review of published cases and clinical recommendations. World J Surg. 2018;42:574-81. https://doi.org/10.1007/s00268-017-4178-1.

[6] Canbay E, Akman E. Appendix perforation in appendix duplication in a man: A case report. J Med Case Rep. 2011;5:01-03. Available from: https://doi.org/10.1186/1752-1947-5-162.

[7] Das R, Cantor JP, Vu TQ. Two concurrent appendiceal neoplasms in an elderly patient: A case report. Int J Surg Case Rep. 2017;40:124-26. Available from: https://doi.org/10.1016/j.ijscr.2017.09.007

[8] Mohamed A, Wu S, Hamid M, Mahipal A, Cjakrabarti S, Bajor D, et al. Management of appendix neuroendocrine neoplasms: Insights on the current guidelines. Cancers. 2022;15(1):295. Available from: https://doi.org/10.3390/cancers15010295.

[9] Jackson M, Marks L, May GH, Wilson JB. The genetic basis of disease. Essays Biochem. 2018;62(5):643-723. Available from: https://doi.org/10.1042/EBC20170053.

[10] Hoehn RS, Rieser CJ, Choudry MH, Melnitchouk N, Hechtman J, Bahary N. Current management of appendiceal neoplasms. Am Soc Clin Oncol Educ Book. 2021;41:118-32. Available from: https://doi.org/10.1200/EDBK_321009.

[11] Shibata J, Tomida A, Hattori M, Hirata A, Imataki H, Orihara Y, et al. Duplication of the appendix masquerading as appendiceal tumour: A case report. Surg Case Rep. 2023;9(1):190. Available from: https://doi.org/10.1186/s40792-023-01769-7.

[12] Duerr EM, Chung DC. Molecular genetics of neuroendocrine tumors. Best Pract Res Clin Endocrinol Metab. 2007;21(1):01-14.

[13] Khan F, Vogel RI, Diep GK, Tuttle TM, Lou E. Prognostic factors for survival in advanced appendiceal cancers. Cancer Biomark. 2016;17(4):457-62. Available from: https://doi.org/ 10.3233/CBM-160662.

[14] Coşkun H, Bostancı Ö, Dilege M, Mihmanlı M, Yılmaz B, Akgün İ, et al. Carcinoid tumours of appendix: Treatment and outcome. Turkish Journal of Trauma and Emergency Surgery. 2006;12(2):150-54.

PARTICULARS OF CONTRIBUTORS:

1. Assistant Professor, Department of Pathology, Osmania Medical College, Hyderabad, Telangana, India.
2. Professor, Department of Pathology, Osmania Medical College, Hyderabad, Telangana, India.
3. Assistant Professor, Department of Pathology, Niloufer Hospital, Hyderabad, Telangana, India.
4. Associate Professor, Department of Pathology, Life Oncopath Laboratories, Hyderabad, Telangana, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. Kovuri Umadevi,
Assistant Professor, Department of Pathology, Osmania Medical College, Hyderabad, Telangana, India.
E-mail: umadeviresearch@gmail.com

PLAGIARISM CHECKING METHODS: [Jain H et al.]

- Plagiarism X-checker: Mar 01, 2025
- Manual Googling: Apr 18, 2025
- iThenticate Software: Apr 28, 2025 (6%)

ETYMOLOGY: Author Origin

EMENDATIONS: 6

AUTHOR DECLARATION:

- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

Date of Submission: **Feb 22, 2025**
Date of Peer Review: **Apr 13, 2025**
Date of Acceptance: **Apr 30, 2025**
Date of Publishing: **Sep 01, 2025**